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# Social life in adult patients after Fontan procedure

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## Abstract

**Background:** Little is known about the social life in adult patients after Fontan palliation. The study aimed to assess the long-term social life of patients after Fontan surgery, including school achievement, employment, and marital status.

**Results:** We conducted a cross-sectional study on patients aged 18 years or above who had a Fontan operation for a single ventricle pathology. Our outcomes were the academic performance and marital and employment status of adult Fontan patients. Patients or their families were interviewed directly or by phone, and a set of questions were asked to address their educational level, employment, and marital status. Ninety-nine patients were included in the study. Their median age was 21 years (min–max: 18–41), and 60% were females ( $n=59$ ). The most common diagnoses were double inlet left ventricle ( $n=24$ , 24%), tricuspid atresia ( $n=21$ , 21%), and the unbalanced atrioventricular canal ( $n=21$ , 21%). Fourteen patients (14%) were married, and 6 of them had children (two were females). Eleven percent were college graduates (either diploma or bachelor's degree), and 47% were high school graduates.

**Conclusions:** Fontan operation could negatively affect the social life of the patients. It may affect employment patterns, educational levels, and marital status.

**Keywords:** Fontan operation, Single-ventricle, Social life

## Background

Fontan procedure is the ultimate pathway for palliation of children with a single ventricle anomaly or its variants. Life expectancy in univentricular heart patients completing the Fontan procedure is relatively good, with a 10-year postoperative survival rate of 91% and 80% at 20 years [1, 2]. However, this procedure remains palliative. Patients are prone to arrhythmias, exercise intolerance, thromboembolic events, heart failure, protein-losing enteropathy, and other severe Fontan-related complications [3, 4].

Studies about the quality of life after Fontan are scarce, and some reports recorded suboptimal quality of life in those patients [5]. Little is known about the impact of Fontan's operation on social life, including school performance, marriage, and employment. It is hypothesized that the Fontan operation could be associated with poor social life in adult patients with univentricular heart physiology. Investigating these long-term social life outcomes will enhance the process of patient and family counseling. Therefore, we aimed to measure the social life outcomes for patients aged 18 years or above who had Fontan palliation during childhood.

## Methods

### Design and population

We conducted a cross-sectional study including patients who had the Fontan procedure. We included patients aged 18 years or above who underwent the Fontan procedure for various cardiac lesions with functionally single

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ventricle during their childhood. We excluded patients with other associated non-cardiac anomalies, neurological anomalies, and associated syndromes, such as Down syndrome.

### Data and outcomes

We collected relevant patient characteristics and operative data from the electronic databases. Our outcomes were adult Fontan patients' school and academic performance, marital, and employment status. Patients or their families (upon patients' request) were directly interviewed or by phone calls between January 2019 and January 2020. A set of questions were asked to address their educational level, employment, and marital status.

### Ethical consideration

This study was conducted per the ethical principles of the Declaration of Helsinki [5]. The Institutional Review Board approval was obtained before study initiation (Reference number: 2181069). As this study is cross-sectional and has no intervention with patients' management, signed consent was waived, and we obtained verbal approval before the patient's enrollment.

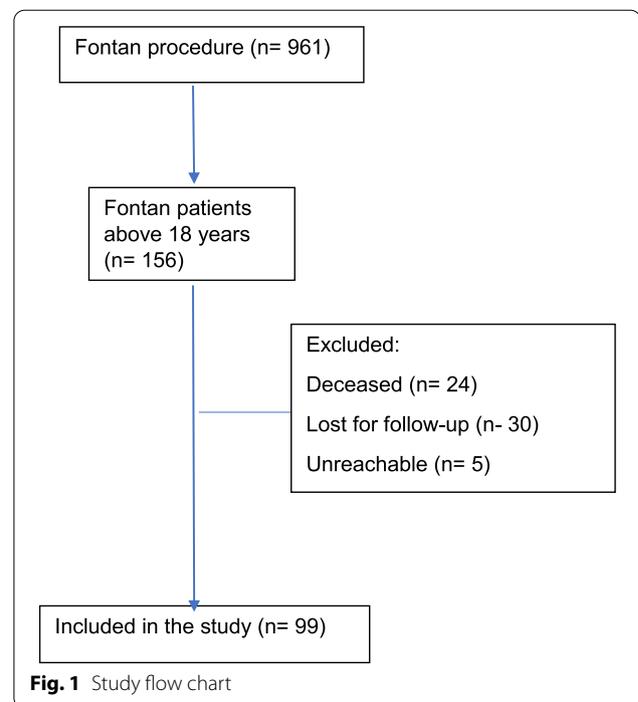
### Statistical analysis

Descriptive statistics were used to present our results. Continuous data were expressed as mean and standard deviation or median and interquartile range, and categorical data as frequencies and percentages.

### Results

A total of 961 Fontan operations patients were identified in our hospital registry. Out of this total number; 156 patients were above 18 years old at the time of our data collection, 24 (15%) were deceased, 30 (18%) lost to follow-up, and 5 (3%) were active but unreachable, ending up with 99 (63%) active and reachable patients to include. The group with lost follow-up had no follow-up between 4 and 18 years (Fig. 1).

Of the ninety-nine patients, 59 were females (60%), and 40 were males (40%). Their age ranged from 18 to 41 years [median: 21 (25th–75th percentiles: 19–25) years]. The median age at the time of Fontan was 4 years (25th–75th percentiles: 3–9 years). Patients were categorized into the following diagnosis categories; double inlet left ventricle (DILV) ( $n= 24$ , 24%), unbalanced atrioventricular septal defect (AVSD) ( $n= 21$ , 21%), tricuspid atresia ( $n= 21$ , 21%), hypoplastic right ventricle, and pulmonary atresia ( $n= 16$ , 16%), hypoplastic left ventricle ( $n= 8$ , 8%), transposition of the great arteries (TGA) and large ventricular septal defect (VSD) ( $n= 8$ , 8%), and Ebstein anomaly ( $n= 1$ , 1%).



Eighty-three were single (83.8%), 14 (14.1%) were married (5 were females), and 2 (2%) were divorced (all were females). Six of the married patients (two of them are females) had children.

Regarding degree achievement, 11% achieved a college degree, 20% are currently in college, 47% finished high school, 6% intermediate school, 8% primary school, and only 5% had no education. Patients who received no education or only primary education had a history of thromboembolic events during or after surgery; some were confirmed by imaging, and others were based on history.

Eighty-four were unemployed, and 15 were employed (13 were males and 2 were females). Twelve patients worked in an office job, while one worked as a nurse, a carpenter, and a security guard.

The oldest Fontan patient included in our study was a 41-year-old male patient. The patient was diagnosed with DILV and TGA. He was a high school graduate, married with four children, and had an office job. The patient recently passed away during the study due to Fontan complications which includes protein-losing enteropathy with chronic ascites, atrial tachycardia, and chronic kidney disease.

We included in our study a 37-year-old male with a DILV. He is a high school graduate, married, and has three children. The patient has an office job, and his general health is satisfactory. Another 31-year-old male with severe Ebstein tricuspid valve disease was included. He

is working as a nurse and is a bachelor's graduate. He is married but does not have children.

We included a 30-year-old female with tricuspid atresia. She successfully delivered three children via c-section. Another 27-year-old female diagnosed with TGA, large VSD, and PA was included. She was 25 weeks pregnant in her 4th pregnancy during the study. She had two miscarriages; her 3rd pregnancy was twin girls born pre-term at 33 weeks old. The mother has a bachelor's degree in business, not employed.

## Discussion

Long-term survival and quality of life are major concerns for parents before Fontan operations [6, 7]. Several studies have explored the quality of life in Fontan patients; however, the results are contradictory [8]. Some reported normal quality of life [9, 10]; others reported reduced quality of life [11, 12]. We performed a study to evaluate the long-term social life of adult patients after the Fontan procedure. The study included 99 adult Fontan patients; the most common diagnoses were DILV, TA, and unbalanced AV canal. Only 31% were in college or achieved a college degree. Fourteen patients were married, five of them were females, and only two of the females had children. Fontan patients attained lower social achievements compared to normal population in our community [13].

Pike and associates compared the quality of life, health status, and prevalence of depression in adolescents or adults after the Fontan procedure to healthy adults [9]. They found that adolescents and adults in the Fontan group reported lower physical health status and were more depressed than their healthy counterparts. There were no differences in quality of life, mental health status, or social support between the two groups.

Pickup and coworkers performed a cohort study to establish the employment complex for adults with congenital heart disease [14]. They included patients with single-ventricle and Fontan circulation. They found the same proportion of professionals in their subjects compared to the rest of the UK. However, there were differences in the job types. Greater proportions of patients were working in the caring, leisure, and other service occupations (15% versus 9%), the elementary occupations (17% versus 11%), sales and customer service occupations (14% versus 8%), and administrative and secretarial occupations (12% versus 11%). The reverse trend was observed for associate professions and technical occupations (7% versus 14%), skilled trades (10% versus 11%), process, plant, and machine operatives (3% versus 6%), and managers, directors, and senior officials (2%

versus 10%). Despite the comparable employment rate, this study showed that the employment pattern is different for single-ventricle patients. The same finding was reported in our series. Fifteen patients were employed; two of them were females. Most of them were working in an office job, which might say about these patients' lower endurance level, making them less capable of withstanding jobs with more physical strain.

Driscoll and associates found preserved fertility after the Fontan operation; however, the abortion rate was high [15]. In our series, two out of five married females were able to have children. We cannot conclude if this number is considered small as our population starts at 18 years, which is not the typical age of marriage, moreover, the marital status is dependent on social circumstances and not only on the medical part.

Several studies demonstrated impaired cognitive, behavioral, and neuropsychological development in Fontan patients [16, 17]. In our study, 31% were college students or achieved a college degree, which is considered low according to the standard age to start college in our country.

## Study limitations

The study has several limitations. First, this is a single-center experience, which limited our cohort's sample size. We included all adult Fontan patients performed at our center and were available to assess the outcomes. Second, we did not include a comparison group. Selection of the comparison group is challenging because of the wide variability of marriage, education, and employment among the average population. Lastly, because of the small sample size, we did not assess the relationship between the baseline and operative characteristics and the study outcome. However, this study highlighted the long-term social outcomes of the Fontan operation in Saudi Arabia and gave an insight into more in-depth parent counseling before surgery.

## Conclusions

Fontan operation could negatively affect the social life of the patients. It may affect employment patterns, educational levels, and marital status.

## Abbreviations

AVSD: Atrioventricular septal; DILV: Double inlet left ventricle; TGA: Transposition of the great arteries; TA: Tricuspid atresia; VSD: Ventricular septal defect.

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**Authors' contributions**

MA: design and drafting; EAA: data collection and drafting; MHA: data collection and data interpretation; FK: research design, supervision, and revision. All authors read and approved the final manuscript.

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**Availability of data and materials**

The authors declare that the data supporting the findings of this study are available upon request

**Declarations****Ethics approval and consent to participate**

King Faisal Specialized Hospital and Research Center, Riyadh, Saudi Arabia (Reference number: 2181069) Patients gave consent before participation

**Consent for publication**

Not applicable.

**Competing interests**

The authors declare that they have no competing interests. Additionally, the authors have no financial interest or ties in or with the product or product distributor.

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